

## LADD PROCEDURE FOR ADULT INTESTINAL MALROTATION: CASE REPORT

*Procedimento de Ladd para má rotação intestinal no adulto: relato de caso*

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### INTRODUCTION

Intestinal malrotation is a rare congenital condition caused by the absence of or incomplete rotation of the small bowel during the embryonic period<sup>1</sup>. The incidence in the general population is one for every 200 to 500 newborns. Symptomatic cases are infrequent, occurring in one to 6000 newborns<sup>1</sup>. The diagnosis is made primarily during the neonatal period, prior to the first month of life. Symptoms are generally acute and small intestine obstruction due to volvulus is observed.

The presentation of intestinal malrotation in adults is rare, and occurs in approximately 0.2%<sup>5</sup>. In general, most adults affected are asymptomatic. Chronic abdominal pain and constipation can occur in some cases. In a review of the literature, there were only 40 cases described in adults between the years 1923 and 1992<sup>5</sup>.

### CASE REPORT

White female with 23 year-old, presented in outpatient colorectal unit with cramping and diffuse abdominal pain beginning during childhood and worsening during the last three months. She reported associated chronic constipation and had utilized stool softeners for the last few years. Bowel movements were infrequent, occurring every three days, with significant effort for defecation. During the last three months, she complained of abdominal distension, infrequent vomiting and worsening of pain and constipation. There was no rectal bleeding or previous conditions treated. There was no previous pregnancy or abdominal operations. The patient complained of constipation since childhood. No

alcohol or tobacco consumption was described.

Physical examination revealed a profile of a thin young female with stable vital signs. Abdominal examination revealed moderate abdominal distension, with significant diffuse pain, without acute peritoneal findings. The general physical examination was normal.

An abdominal ultrasound study and a full colonoscopy were performed and revealed no abnormal findings. A small bowel contrast imaging study revealed a duodenal rotation defect, with the whole small intestine (jejunum and ileum) on the right side of the midline and a vertical duodenum, without duodenojejunal angle (Figures 1 and 2). Despite the described rotation abnormalities, the barium was normally processed through the right colon, located in front of the lumbar spine, next to the left lobe of the liver. MRI showed normal positioning of the superior mesenteric vessels.



FIGURE 1 – Small bowel imaging showing a vertical duodenum and absence of duodenojejunal angle

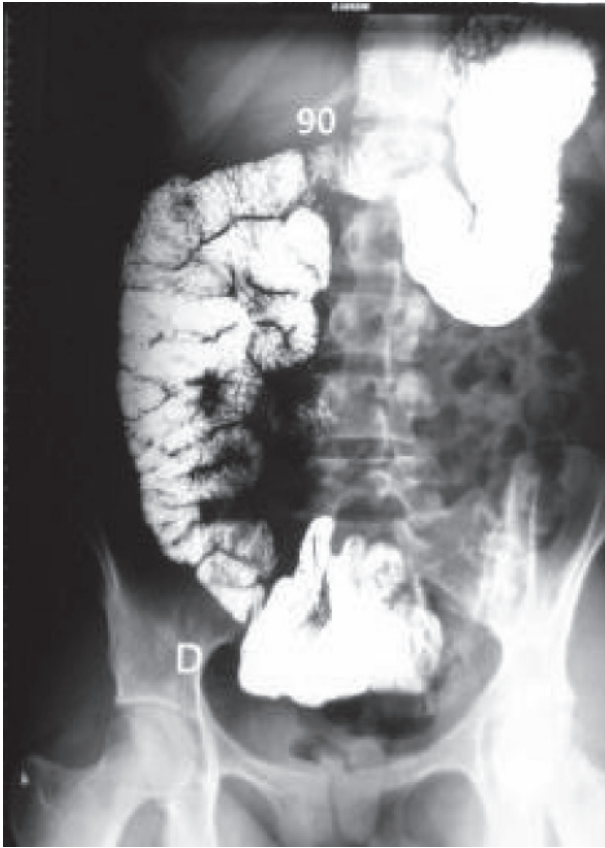


FIGURE 2 – Small bowel imaging with the entire small bowel on the right side of the abdomen

With the diagnosis of symptomatic intestinal malrotation, the patient underwent laparotomy and Ladd procedure. After abdominal cavity inspection, the most important findings included the absence of duodenojejunal angle and the 3<sup>rd</sup> and 4<sup>th</sup> duodenal segments spreading downward to the right iliac fossa. The entire small bowel was located on the right side of the abdomen, and the colon was fixed in the left quadrants. There were multiple adhesions from the duodenum to the peritoneum of the right upper quadrant (Ladd's adhesions), that were submitted to sharp dissection. The small bowel was released and placed in the normal anatomic position, without any fixation. Appendectomy was also performed.

The patient responded well during the post-operative period. She had no abdominal cramps and constipation decreased significantly. She remains asymptomatic two years after surgery.

This report was approved by the Ethical Board of Cajuru University Hospital. The patient signed an informed consent authorizing this publication.

## DISCUSSION

Congenital abnormalities of intestinal rotation are rarely found in adults<sup>1</sup>. Most of the cases are diagnosed

during the neonatal period. Intestinal malrotation can lead to unspecific chronic symptoms in young adults, and are difficult to diagnose. The knowledge of intestinal embryology is essential for the comprehension of the mechanism of its formation<sup>2</sup>.

The absence of a complete rotation of the midgut, during the embryony period, is the key to the physiopathology of intestinal malrotation. The duodenum does not assume its normal position, posterior to the superior mesenteric artery. Consequently, there is no fixation of the mesentery in the posterior abdominal wall. This causes intestinal torsion through the superior mesenteric artery, one of the most common complications of rotation abnormalities<sup>2</sup>.

Intestinal torsion anomalies can be classified according to the stage of occurrence. Stage 1 is mainly the onphalocele; stage 2 includes malrotation, non-rotation and reverse rotation of the gut; stage 3 anomalies include mobile cecum, mobile duodenum and free mesentery of the small bowel<sup>1</sup>.

Clinical presentation of intestinal malrotation can be unspecific. Dietz *et al.*, in 2002, described the Cleveland Clinic series of 10 adults with this form of intestinal anomaly<sup>2</sup>. They described two different groups of patients, according to their symptoms. The first group of patients presented chronic symptoms of bowel obstruction characterized by a history of recurring episodes of nausea, bilious vomiting, and abdominal cramping and pain. Abdominal distention was usually not a feature as the obstruction tended to be proximally located. Constipation can be a common symptom in these cases. The second group had symptoms related to the acute onset of obstruction, mainly due to adhesion of the Ladd's bands or volvulus. In this case, the patient had intermittent constipation and mild abdominal pain, and can be included in the first group previously described.

Most of intestinal malrotation patients present signs of obstruction during neonatal period, and this condition should be considered in all newborns with bilious vomiting and abdominal pain<sup>1</sup>. Older children and adults more often present recurrent mild symptoms, as did this patient. Approximately 30% of these patients present vomiting and 20% have recurrent unspecific abdominal pain<sup>1</sup>. The differential diagnosis includes other causes of vomiting and pain, including peptic ulcers, functional disorders, irritable bowel syndrome or psychiatric disorders<sup>1,5</sup>.

The diagnosis of intestinal malrotation can be confirmed with upper GI tract contrast imaging<sup>3,4</sup> that can reveal a vertical duodenum, with a right location in the abdominal cavity, and the absence of the duodenojejunal angle. These results are found in nearly 80% of the patients<sup>4</sup>. However, sometimes this study can be normal. A double-contrast barium enema can show the abnormal cecal location, just below the liver, near the midline, and the entire colon located laterally

to the spine on the left side<sup>3</sup>. CT scan can also identify these abnormal positions of the small bowel and the colon and the opposite positioning of the superior mesenteric vein, located on the left side of the artery. It may also be helpful in identifying acute obstruction<sup>3,4</sup>. In the reported case, the diagnosis was confirmed with a simple small intestine contrast imaging.

The Ladd procedure, initially described in 1936, is the classic surgical treatment for intestinal malrotation<sup>1</sup>. It is described as an association of the mobilization of the duodenum and the right colon, section of the Ladd's bands, section of possible adhesions near the superior mesenteric vessels and appendectomy. The aim of this procedure is to reduce the risk of acute volvulus, by locating the small intestine in a non-rotating position and widening the base of the mesentery. Appendectomy is performed due to possible difficulty in the diagnosis of future appendicitis, distant from the classic lower right quadrant position.

The Ladd procedure can also be safe if performed by laparoscopy. Although most reports describe laparotomy as a classic approach, more recent studies point out the efficacy and feasibility of laparoscopy<sup>5,6</sup>. Mazziotti *et al.* described a method to assure the possibility of laparoscopic Ladd procedure, on every occasion that the length between the duodenojejunal junction and the ileocecal valve is less than half the transverse diameter of the peritoneal cavity<sup>6</sup>.

The Mayo Clinic described a series of 21 adults with intestinal malrotation, treated with Ladd procedure.

Eleven of these patients underwent laparoscopic surgery, and were compared with ten patients operated by laparotomy. They concluded that laparoscopy is as safe, feasible and efficient as the open procedure, with all the advantages of this method related to early discharge and oral intake<sup>5</sup>.

In the present case, laparotomy was performed due to lack of experience by our team in managing this rare condition.

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## ERRATA

In the edition of volume 22 (1): 61 there was an error in the abstract of the article. ABCD apologizes to authors and makes the correction.

**ABSTRACT – Background** – The differential diagnosis of the unconscious patient must always include hyperosmolar hyperglycemic non-ketotic hypothesis. **Case report** – A 22 year-old woman, ABO – O+ with history of fatigability and jaundice. Physical examination revealed a markedly jaundice patient. Fulminant hepatic failure was the diagnostic. Liver transplant was performed from a brain- dead cadaver donor with success. Arterial hepatic thrombosis was considered one week after liver transplant and confirmed with Doppler-US. The hepatic retransplant occurred without problems. After two days of liver transplant the serum glucose was 600 mg/dl and unconsciousness. Hyperosmolar coma was controlled and treated with succes for 48 h. The patient left the hospital after 30 days of liver transplantation without diabetes. **Conclusion** – Hyperosmolar coma is an rare event after liver transplant. The early recognition and treatment of this disorder should result in improvement of evolution.

**HEADINGS** – Liver transplantation. Hyperosmolar. Hyperglycemic.